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CASE REPORT

A case of persistent hemifacial weakness

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FIGURE 1
A 33-year-old woman presented with facial weakness and synkinesis (left). Notice the involuntary eye closure, poor smile and abnormal contraction of the cheek and chin muscles. Patients frequently complain of a cramp-like sensation due to constant muscle activity. Following facial neuromuscular retraining and chemodenervation of aberrant muscle activity her facial function was much improved (right)

What are the causes of facial palsy?

HEMIFACIAL WEAKNESS, OR PALSY, RESULTS FROM DISRUPTION OF COMMUNICATION BETWEEN cortical motor centres and the facial musculature along the course of the facial nerve. Facial palsy is not an uncommon presentation in primary care and can present both diagnostic challenges as well as physical and psychosocial problems for patients.

Our patient, see box 1, p30, highlights a typical presentation of Bell's palsy — sudden onset, mild otalgia, altered facial sensation and/or taste, with no obvious prodrome.[1] A lower motor neurone palsy of unknown aetiology, more prevalent during pregnancy and in those with a family history,[2] it represents over half of hemifacial weakness cases in primary care. However, as a diagnosis of exclusion, there are a number of key clinical features of more sinister diagnoses that must be considered see table 1, p30.

Which patients should be referred?

Acute hemifacial weakness secondary to isolated facial nerve palsy must be differentiated from acute cerebrovascular accident (CVA). The latter results in sparing of the brow musculature due to the bilateral innervation of the frontalis. Bell's palsy is also not associated with the other corporeal neurological deficits of CVA. Altered facial sensation and mild otalgia are common in Bell's palsy, however severe pain is suggestive of Ramsay Hunt syndrome, with pain frequently present before the emergence of vesicles. Ramsay Hunt syndrome may also be associated with sudden onset vestibulocochlear nerve involvement (tinnitus, vertigo, and sensorineural hearing loss) as well as intraoral pain.[3] Recent facial or head trauma and surgery should also be excluded in causation.

What are the management options?

Facial palsy secondary to tumours of the head and neck is generally of slow, progressive onset and is almost always preceded by other symptoms. Specifically, tumours of the »

Box 1

Case history

A fit and well 33-year-old woman presented to her GP with a 12-hour history of right-sided facial weakness and ear pain. Examination confirmed right-sided facial weakness with nonspecific sensory changes and lagophthalmos. The patient was systemically well, showed no other neurological signs, and otological examination was normal.

The case history represents a typical presentation of Bell's palsy. Unfortunately, unlike the majority of patients, her facial palsy and lagophthalmos persisted; over time she developed distressing synkinesis (involuntary facial twitching/spasms), depression, anxiety, and headaches.

Like many sufferers there was a long interval between the onset of facial palsy and referral as it was felt her condition was primarily cosmetic and the synkinesis was an inevitable (untreatable) sequela of the condition. She was referred to a specialist service where directed physical therapy and botulinum toxin injections controlled aberrant synkinetic movements to restore her facial function and improve her quality of life. Figure 1, p29, demonstrates what can be achieved with targeted facial exercises.

cerebellopontine angle e.g. acoustic neuromas, present with tinnitus and hearing loss, and parotid tumours with a mass.[4]

'In the assessment of acute facial palsy examination should include the cranial nerves, ear, parotid, neck and oral cavity'

Middle and external ear infections do not cause facial palsy in isolation. Significant otitis externa in a patient with diabetes may predate the development of facial palsy, as would progressive hearing loss and the foul persistent discharge of cholesteatoma. Facial palsy is seen in persistent acute otitis media however it is a rare complication.

Facial palsy may also be associated with systemic disease such as sarcoidosis or HIV and should be considered within the spectrum of concurrent symptoms and medical history. The exception is that of Lyme neuroborreliosis where facial palsy is occasionally the sole presenting symptom.[5]

REFERRAL

Acute Bell's palsy can be managed successfully in primary care. Current research still demonstrates benefit in the expeditious use of steroids (e.g. 50 mg prednisolone once daily within 72 hours of onset for ten days). However no clear added benefit from the use of antiviral therapy has been shown.[1]

In the assessment of acute facial palsy examination should include the cranial nerves, ear, parotid, neck and oral cavity. It is essential for both GPs and specialists to grade the extent of weakness early to determine prognosis and direct treatment, see table 2, opposite.

The following patients should be referred:

- Cases with lagophthalmos require eye protection (patching and artificial tears); ophthalmological opinion is required if the weakness persists beyond a few days or ocular damage is suspected
- In Ramsay Hunt syndrome, which typically presents with severe pain and a rash on the ipsilateral ear or intraorally, antivirals both expedite recovery and improve outcome.[3] The usual dose is aciclovir 400 mg five times daily for ten days. Referral to specialist care is indicated in immunocompromised patients, those with significant pain, and where intraoral vesicles prohibit oral intake
- Palsy secondary to trauma or surgery should be referred immediately as surgical exploration is often required

Table 1

Red flags and diagnoses in facial palsy

Red flag	Cause	Potential diagnoses
Forehead sparing with other acute neurology	Upper motor neurone sparing	Cerebrovascular event
Parotid mass	-	Parotid tumour
Hearing loss, tinnitus, vertigo	Associated compression of vestibulocochlear nerve	Cerebellopontine angle tumour
Otitis externa in a diabetic patient	Erosion of external auditory canal	Malignant otitis externa
Trauma (or recent surgery)	-	Trauma
Acute severe or chronic otitis media	Middle ear or mastoid disease	Otitis media +/- cholesteatoma
Bilateral facial weakness	-	Sarcoidosis, Guillain-Barré syndrome
Systemic upset	-	Ramsay Hunt syndrome, HIV, leukaemia, sarcoidosis, Lyme disease
Herpetic vesicles/severe pain	Herpes simplex virus in geniculate ganglion	Ramsay Hunt syndrome
Other significant neurology	-	CVA, intracranial tumour, multiple sclerosis
Tick bite (or exposure) Erythema chronicum migrans	-	Lyme disease (neuroborreliosis)

Table 2

House-Brackmann grading of facial palsy

Grade	Description	Characteristics
I	Normal	Normal facial function in all areas
II	Mild dysfunction	Slight weakness noticeable on close inspection; may have very slight synkinesis
III	Moderate dysfunction	Obvious, but not disfiguring, difference between two sides; noticeable, but not severe, synkinesis (involuntary facial twitching/spasms), contracture, or hemifacial spasm; complete eye closure with effort
IV	Moderately severe dysfunction	Obvious weakness or disfiguring asymmetry; normal symmetry and tone at rest; incomplete eye closure
V	Severe dysfunction	Only barely perceptible motion; asymmetry at rest
VI	Total paralysis	No movement

Table 3

Management of persistent facial palsy

Problem	Therapy/specialist
All aspects of facial weakness and psychological support/advice	Facial rehabilitation Specialist physiotherapy Speech and language therapy
Synkinesis/twitching (facial hyperactivity)	Specialist physiotherapy Plastic surgery — botulinum toxin Acupuncture
Dry eyes, recurrent abrasion, infections	Eye care, ophthalmology
Speech problems	Speech and language therapy
Poor dental hygiene and dry mouth	Dentist
Psychological problems	Psychology/psychiatry
Asymmetrical smile/incomplete eye closure/facial asymmetry	Plastic surgery
Advocacy and support	Facial Palsy (UK) www.facialpalsy.org.uk

- A parotid mass with facial palsy implies malignant change and must be referred within the two-week wait pathway
- A history of progressive hearing loss and tinnitus with palsy also requires urgent referral to neuro-otology for assessment of cerebellopontine angle tumours
- All cases of facial palsy associated with infective otological symptoms should be discussed with ENT

MANAGEMENT

If an organic cause is excluded, reassurance and advice are critical when managing facial weakness. Prognosis is dependent upon causation.

Bell's palsy results in full recovery in around 70% of patients,[1] partial paralysis is prognostically favourable. Recovery is usually rapid (within three weeks) but can take up to 12 months. There is recurrence in around 10% of cases.[2] The longer the duration of paralysis in Bell's palsy the poorer the recovery. Episodes occurring during pregnancy have a slightly worse outcome.

If no recovery is observed after six weeks, a referral to an ENT or neurology department is appropriate as exclusion of a space-occupying lesion is important.

Recovery in Ramsay Hunt syndrome is less complete with only 60% returning to House-Brackmann grade I or II. Incomplete palsy resulting from trauma or surgery usually resolves completely. However, recovery following complete traumatic palsy has a more variable outcome.

'It is essential for both GPs and specialists to grade the extent of weakness early to determine prognosis and direct treatment'

Preoperative weakness associated with tumours is often irreversible because compromised nerve is sacrificed where resection is necessary. However, postoperative impairment frequently recovers after resolution of transient oedema.

More complex than acute management is the long-term therapy of patients with facial nerve palsy. Weakness is not the only symptom requiring therapeutic intervention. Varying degrees of palsy also affect salivation, dental care, speech, eating, eye closure, and responsive facial animation.

Facial palsy is commonly associated with symptoms of anxiety and/or depression, often poorly correlated with the degree of weakness, and frequently overlooked.[6] Thus, a multidisciplinary assessment to rehabilitate and manage the functional and aesthetic as well as the psychological aspects is essential (see table 3, above). Facial therapists formulate strategies to aid symptom optimisation, including neuromuscular training and physiotherapy that may aid those patients with chronic symptoms.[7]

Selected patients may also then benefit from a range of reconstructive surgical treatments. »

CASE REPORT

HEMIFACIAL WEAKNESS

key points

SELECTED BY

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Hemifacial weakness, or palsy, results from disruption of communication between cortical motor centres and the facial musculature along the course of the facial nerve. Bell's palsy has a typical presentation of sudden onset, mild otalgia, altered facial sensation and/or taste, with no obvious prodrome. It represents over half of hemifacial weakness cases in primary care. However, as a diagnosis of exclusion, there are a number of key clinical features of more sinister diagnoses that must be considered.

Acute hemifacial weakness secondary to isolated facial nerve palsy must be differentiated from acute cerebrovascular accident. The latter results in sparing of the brow musculature due to the bilateral innervation of the frontalis. Altered facial sensation and mild otalgia are common in Bell's palsy, however severe pain is suggestive of Ramsay Hunt syndrome. Recent facial or head trauma and surgery should also be excluded in causation.

Facial palsy secondary to tumours of the head and neck is generally of slow, progressive onset and is almost always preceded by other symptoms. Middle and external ear infections do not cause facial palsy in isolation. Facial palsy is seen in persistent acute otitis media however it is a rare complication. It may also be associated with systemic disease such as sarcoidosis or HIV.

Acute Bell's palsy can be managed with the expeditious use of steroids (e.g. 50 mg prednisolone once daily within 72 hours of onset for ten days), however there is no clear added benefit from the use of antiviral therapy (as there is in Ramsay Hunt syndrome.)

Patients with the following conditions should be referred: lagophthalmos if the weakness persists beyond a few days or ocular damage is suspected; Ramsay Hunt syndrome (immunocompromised patients, those with significant pain, and where intraoral vesicles prohibit oral intake); and palsy secondary to trauma or surgery. A parotid mass with facial palsy implies malignant change and must be referred within the two-week wait pathway. A history of progressive hearing loss and tinnitus with palsy also requires urgent referral to neuro-otology for assessment of cerebellopontine angle tumours. All cases of facial palsy associated with infective otological symptoms should be discussed with ENT.

Bell's palsy results in full recovery in around 70% of patients, partial paralysis is prognostically favourable. Recovery is usually rapid (within three weeks) but can take up to 12 months. There is recurrence in around 10% of cases. The longer the duration of paralysis in Bell's palsy the poorer the recovery. Recovery in Ramsay Hunt syndrome is less complete with only 60% returning to House-Brackmann grade I or II.

Facial palsy is commonly associated with symptoms of anxiety and/or depression, often poorly correlated with the degree of weakness, and frequently overlooked. Facial therapists formulate strategies to aid symptom optimisation, including neuromuscular training and physiotherapy that may aid those patients with chronic symptoms.

CONCLUSIONS

Acute facial weakness frequently presents a diagnostic dilemma in general practice. If in doubt, patients should be discussed with secondary care. However, most can be managed successfully in primary care with the likelihood of a good recovery.

'Facial palsy is commonly associated with symptoms of anxiety and/or depression, often poorly correlated with the degree of weakness, and frequently overlooked'

Facial palsy is not just a cosmetic problem, but rather a condition associated with long-term functional and psychological sequelae for which a range of effective treatment options are available. Advice and support is essential. Charities and support groups are invaluable in the education of patients and their families (see Useful information box, below).

REFERENCES

1 Gronseth GS, Paduga R. Evidence-based guideline update: Steroids and antivirals for Bell's palsy. Report of the Guideline Development Subcommittee of the American Academy of Neurology. *Neurology* 2012; 79(22):2209-13
2 Oosterveer DM, Benit CP, deSchryver E. Differential diagnosis of recurrent or bilateral peripheral facial palsy. *J Laryngol Otol* 2012;126:833-836
3 Ryu EW, Lee H, Lee SY et al. Clinical manifestations and prognosis of patients with Ramsay Hunt syndrome. *Am J Otolaryngol* 2012;33(3): 313-8
4 Stodulski D, Mikaszewski B, Stankiewicz C. Signs and symptoms of parotid gland carcinoma and their prognostic value. *Int J Oral Maxillofac Surg* 2012;41(7): 801-6
5 Broekhuijsen-van Henten DM, Braun KP, Wolfs TF. Clinical presentation of childhood neuroborreliosis; neurological examination may be normal. *Arch Dis Child* 2010 Nov; 95(11):910-4. Epub 2010 Jun 28
6 Walker DT, Hallam MJ, Ni Mhurchadha S et al. The psychosocial impact of facial palsy: our experience in one hundred and twenty six patients. *Clin Otolaryngol* 2012;37(6:474-7.doi:10.1111/coa.12026
7 Teixeira LJ, Valbuza JS, Prado GF. Physical therapy for Bell's palsy (idiopathic facial paralysis) *Cochrane Database Syst Rev* 2011 Dec 7;(12) CD006283

Useful information

Facial Palsy UK
The charity provides support and advice for patients as well as information for healthcare professionals
www.facialpalsy.org.uk

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